Pallidal hyperintensities – a coincidental finding of clinical relevance in Miller Fisher syndrome

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Keywords: brain MRI, hepatic ultrasound, Miller Fisher syndrome, pallidal hyperintensity, porto-systemic shunt

Received 15 December 2010
Accepted 22 February 2011

Sir,

A 41-year-old healthy woman presented with a 3-day history of dysarthria, diplopia and vertigo, preceded by a common cold 2 weeks earlier. Neurological examination showed a nearly complete ophthalmoplegia, truncal ataxia and lower limb areflexia. Miller Fisher Syndrome (MFS) was suspected based on clinical presentation, electrophysiological studies and the presence of antibodies against GQ1b. Brain MRI, performed as part of the initial work-up, unexpectedly demonstrated bilateral T1 pallidal hyperintensities (Fig. 1a). This was considered as unrelated to MFS, a variant of Guillain–Barré syndrome, in which brain MRI is usually normal, with the exception of very scarce reports on high intensity abnormalities in the brainstem or cranial nerves [1].

Differential diagnosis of T1 pallidal hyperintensities included manganese accumulation, nonketotic hyperglycemia [2], renal failure, multiple system atrophy [3] and cerebral anoxia [4]. All of them were excluded in our patient except manganese accumulation occurring secondary to liver failure or professional exposure (welders for example), and reported to produce isolated T1 pallidal hyperintensity in patients with parkinsonism, gait ataxia, encephalopathy or tremor [5]. Our patient did not have any tonus changes, memory loss, or tremor, and the biological hepatic work-up was normal. However, because of the MRI findings, a hepatic ultrasound was carried out and revealed an intra-hepatic porto-systemic shunt (Fig. 1b).

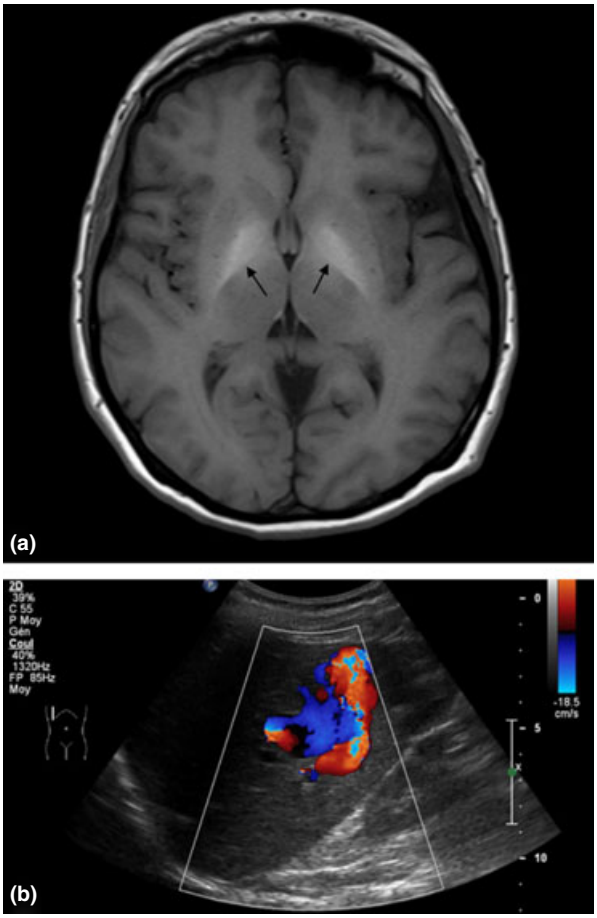

Figure 1 (a) Cerebral MRI showing marked symmetrical pallidal hyperintensities (arrows) on T1 sequences. (b) Hepatic color-coded Doppler ultrasound demonstrating a massive intrahepatic portosystemic shunt.

At six months, the patient recovered completely and at the time of manuscript submission, 10 months later, she was in good health and had no abnormal signs. The hepatic shunt was not closed because of the inherent risk of such intervention in an asymptomatic patient. Brain MRI was not repeated because of good clinical outcome.

This case underlines the need to have good correlations between neuroimaging findings and clinical picture: T1 pallidal hyperintensities are clearly not associated with MFS and in undetermined cause of T1 pallidal hyperintensities a hepatic and metabolic work-up is warranted even in the absence of parkinsonism or cognitive impairment.

Disclosure of conflict of interest

Dr. Thierry Kuntzer serves on editorial boards of Journal of the Peripheral Nervous System, and of Neurophysiologie clinique/Clinical Neurophysiology, and on scientific boards of Société Francaise de Myologie and of Société Francophone du Nerf Périphérique. Dr. Francesca Siclari and Dr. Vincent Alvarez report no disclosures.

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